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Case report

Adenoid cystic carcinoma of the external auditory canal associated with cholesteatoma in an 8-year-old girl

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ABSTRACT

Adenoid cystic carcinoma of the external auditory canal is very rare in a child, and has not previously been reported in an individual younger than 18 years. We report an 8-year-old girl with adenoid cystic carcinoma of the external auditory canal associated with cholesteatoma. At the time of diagnosis, the tumor had invaded the surrounding structures and metastasized to the lymph nodes. She underwent palliative surgery and radiotherapy. She subsequently deteriorated and died of her disease 31 months later.

1. Introduction

Adenoid cystic carcinoma (ACC) is very rare in a child, especially in the external auditory canal (EAC) [1–3]. The histopathological features of ACC in the EAC are similar to those of ACC at other sites. The typical histological patterns are tubular, cribriform, and solid. Perineural invasion is common, but lymph node metastasis is rare. The most common symptoms are otalgia, otorrhea, and a mass in the EAC. The treatment of choice is surgical excision followed by radiotherapy. Cervical lymph node dissection is not recommended in patients without demonstrated lymph node metastasis. Chemotherapy is not recommended because of inconsistent results.

2. Case report

An 8-year-old girl with a 6-month history of left-sided otalgia was assessed at her local hospital. She was diagnosed with cholesteatoma, and underwent canal wall up tympanomastoidectomy. Pathological examination of the surgical specimen confirmed cholesteatoma. Forty days after her operation, otalgia recurred, and at 4 months after her initial operation she underwent further surgery. Her otalgia continued to worsen, and 12 months after her initial operation she was referred to our hospital because of otorrhea and left facial palsy. Physical examination revealed a gray-red tumor in the left external auditory canal and enlargement of left cervical lymph nodes. Computed tomography and magnetic resonance imaging showed a malignant tumor in the EAC (Fig. 1). Histological examination of a biopsy specimen showed adenoid cystic carcinoma. Because of extensive tumor invasion, we performed palliative surgery (subtotal resection of the temporal bone with positive surgical margins including dura mater of the baldaquin and jugomaxillary muscles, and selective dissection of neck zones II and III), followed by radiotherapy.

Twelve months after resection, the patient experienced left-sided headaches, followed by left-sided hearing loss, loss of sight in the left eye, and bilateral lower limb paralysis. Eighteen months after resection, she died of her disease.

Pathological examination of the surgical specimen showed tubular and cribriform adenoid cystic carcinoma with invasion of the adjacent nerves, parotid gland, skeletal muscles, bone, and cartilage (Fig. 2). Three of the 10 harvested lymph nodes had metastatic disease. Immunohistochemical staining of myoepithelial cells was positive for SMA and p63, and staining of glandular cells was positive for CK7. All glandular cells and 50% of the myoepithelial cells stained positive for CD117. The Ki-67 index was 20%. There were no mutations of exons 9, 11, 13, or 17 of the c-kit gene.

3. Discussion

Only 20% of EAC tumors are glandular in origin, of which ACC is the most common [1,2]. ACC of the EAC has similar features to ACC

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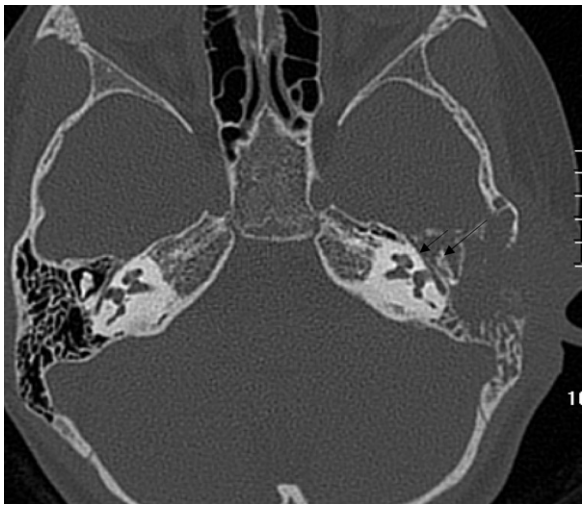

Fig. 1. High-resolution computed tomography. Axial section showing bone destruction in the squamous and mastoid parts of the left temporal bone, disruption of the lateral wall of the tympanic segment of the facial nerve canal (short arrow), and decreased density of the auditory ossicles (long arrow) indicating tumor invasion.

of the salivary gland, with slow growth, high local recurrence rate, perineural invasion, and late distal metastasis. The incidence of ACC of the EAC peaks in the 5th and 6th decades of life, with the youngest patient reported to date being 19 years old [3]. Symptoms include otalgia, otorrhea, and an EAC mass, and are usually present for several years before diagnosis [1,3]. The treatment of choice is complete resection followed by radiotherapy. Chemotherapy is not recommended. As the addition of postoperative radiotherapy results in significantly better local control than surgery alone, some authors recommend treatment with radiotherapy only in patients with advanced tumors [1]. In this case, we performed palliative surgery and postoperative radiotherapy because of extensive tumor invasion. The patient's condition deteriorated soon after treatment, suggesting that radiotherapy only would not have been very effective. Distal metastasis is commonly detected 5 years or longer after diagnosis. The most common sites of distal metastasis are the lungs, kidneys, and vertebrae. The bilateral lower limb paralysis in our patient suggests vertebral metastasis, which was unfortunately not confirmed by imaging.

Our patient was only 8 years old at the time of diagnosis. She had a 6-month history of otalgia before being diagnosed and treated for cholesteatoma. She then deteriorated for another year before being admitted to our hospital, by which time there was extensive tumor invasion into adjacent structures and it was too late for curative resection. Otalgia and otorrhea are common symptoms, as are symptoms related to other nearby structures such as rhinitis, nasopharyngitis, and odontalgia. In the early stages, tumors of the EAC may only be visible as mucosal swelling rather than a mass. When two lesions occur simultaneously, the less visible lesion may initially be missed. If a patient's symptoms cannot be explained by a common lesion, or the symptoms get worse after initial treatment for a common lesion, a diagnosis of tumor should be considered, even if the patient is a young child. Appropriate imaging and biopsy should then be performed to exclude tumor. Dong et al. found that patients who were symptomatic for less than 2 years before diagnosis had a better prognosis than those who were symptomatic for a longer period [3]. In this case, the patient was only symptomatic for 18 months before diagnosis with extensive invasion, and it was 49 months from the onset of symptoms until death, which suggests that ACC is more aggressive in children than in adults.

Lymph node metastasis occurs in approximately 4–5% of patients with ACC [1,4]. Cervical lymph node dissection is not routinely performed unless positive nodes are demonstrated [1,4]. In this case, metastasis was found in 3 of 10 harvested lymph nodes. Although ACC progresses slowly in the early stage and early lymph node metastasis is rare, node metastasis may be more common in the late stage, and such patients should be investigated

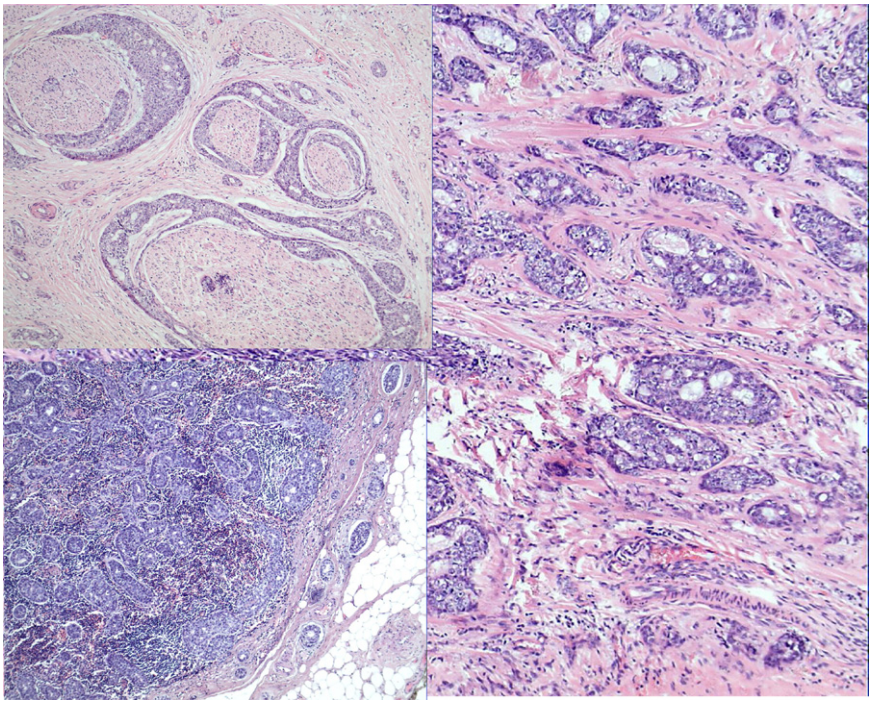

Fig. 2. The tubular and cribriform cellular pattern of classical ACC. Note the perineural invasion (upper left) and lymph node metastasis (lower left).

for metastasis to cervical lymph nodes as well as lung and bone. Our patient died 30 months after treatment, indicating that lymph node metastasis may be associated with poor prognosis. The occurrence of possible bone metastasis and definite lymph node metastasis during such a short clinical course suggests that ACC may be more aggressive in children than in adults, and that routine lymph node examination is important.

The etiology of ACC remains unclear. Most cases show overexpression of CD117, the protein product of the c-kit gene [4]. Recent research has therefore focused on mutations in the c-kit gene. If a mutation is found, targeted therapy may be possible, especially in patients with advanced disease. However, the results of such therapy are still unclear. The estimated prevalence of c-kit gene mutation in patients with ACC ranges from rare to 87.5% [5–7]. In this case, CD117 overexpression was observed, but no c-kit gene mutation was found. It may be that c-kit gene mutation does not play an important role in the pathogenesis of ACC.

4. Conclusions

Adenoid cystic carcinoma of the EAC is very rare in children, and is more aggressive than in adults. Lymph node and distal metastasis may occur, and the prognosis is poor. Prompt and complete imaging examination and biopsy are helpful for early detection, leading to earlier treatment to improve survival and quality of life for children with this condition.

Conflict of interest statement

The authors declare no conflict of interest.

Consent

Written informed consent was obtained from the patient's parents for publication of this case report and any accompanying images.

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